Maryland
DEPARTMENT OF HEALTH

Brineura (cerliponase alfa)

Brineura is a hydrolytic lysosomal N-terminal tripeptidyl peptidase indicated to slow the loss of ambulation in symptomatic pediatric patients 3 years of age and older with Late Infantile Neuronal Ceroid Lipofuscinosis type 2 (CLN2), also known as Tripeptidyl Peptidase 1 (TPP1) Deficiency.

I. Criteria for Initial Approval

Brineura will be considered for coverage when **all** of the criteria below are met, confirmed with supporting medical documentation.

- Patient is 3 years of age and older.
- Prescribed by a neurologist or specialist in this disorder.
- Documentation of a clinical diagnosis of Late Infantile Neuronal Ceroid Lipofuscinosis type 2 (CLN2); Tripeptidyl Peptidase 1 (TPP1) Deficiency.
    - Patients must have a definitive diagnosis of late infantile CLN2 confirmed by deficiency of the lysosomal enzyme tripeptidyl peptidase-1 (TPP1) and/or molecular analysis indicating dysfunctional mutation of the TPP1 gene on chromosome 11p15.
    - Patient has mild to moderate disease documented by a two-domain score of 3-6 on motor and language domains of the Hamburg CLN2 Clinical Rating Scale, with a score of at least 1 in each of these two domains.
    - Patient is ambulatory. Medication must be used to slow the loss of ambulation in symptomatic patients.
- Patients cannot have any of the following contraindications to therapy:
    - Acute intraventricular access device-related complications (e.g., leakage, device failure, or device-related infection).
    - Ventriculoperitoneal shunts.
    - No signs or symptoms of acute, unresolved localized infection on or around the device insertion site (e.g. cellulitis or abscess); or a suspected or confirmed central nervous system (CNS) infection (e.g., cloudy CSF or positive CSF gram stain, or meningitis).

- Patients with a history of bradycardia, conduction disorder, or with structural heart disease must have electrocardiogram (ECG) monitoring performed during the infusion.

II. Criteria for Continuation of Therapy

All of the criteria for initial therapy (in **Section I.**) must be met; **AND**
The provider must attest to a positive clinical response.

- Documentation of at least a 1 in the Motor domain of the CLN2 Clinical Rating Scale.

III. Dosing/Administration

Brineura must be administered according to the current FDA labeling guidelines for dosage and timing. The recommended dosing is as follows:

- 300 mg administered once every other week as an intraventricular infusion followed by an infusion of Intraventricular Electrolytes over approximately 4.5 hours.

IV. Length of Authorization for Initial Therapy

Brineura will be authorized for 6 months when criteria for initial approval are met. Continuing therapy with Brineura will be authorized for 12 months.

V. Billing Code/Information

J0567 Injection, cerliponase alfa, 1 mg;1 billable unit = 1 mg.

Prior authorization of benefits is not the practice of medicine nor the substitute for the independent medical judgment of a treating medical provider. The materials provided are a component used to assist in making coverage decisions and administering benefits. Prior authorization does not constitute a contract or guarantee regarding member eligibility or payment. Prior authorization criteria are established based on a collaborative effort using input from the current medical literature and based on evidence available at the time.

Approved by MDH Clinical Criteria Committee: 2/23/2021
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